INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

XANTHOMA DISSEMINATUM: A RARE NON-LANGERHANS CELL HISTIOCYTOSIS

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Background: A 76-year-old male has a 1.5-year history of an asymptomatic dermatitis in the setting of a macrocytic anemia and thrombocytopenia. A previous biopsy reported leukocytoclastic vasculitis and a dense neutrophilic infiltrate; direct immunofluorescence was negative. A bone marrow aspirate (BMA), colonoscopy and CT scans found no abnormalities.

Observation: He had several infiltrated yellow plaques on the neck, face, palms and axillae. Histopathology revealed a dermal infiltrate with foamy histiocytes (S-100-, CD68+, Factor XIIIa+), lymphocytes, neutrophils, leukocytoclasia, and necrobiosis. Histiocytic infiltrate showed phagocytosis without emperipolesis or cytophagocytosis. Gram, Giemsa, PAS-D, GMS, AFB, Hale's coloidal iron, and Prussian blue stains were negative. Xanthoma disseminatum was diagnosed. Flow cytometry was unremarkable. High LDH was observed. BMA reported a myelodysplastic syndrome with single lineage dysplasia in erythrocytes. Treatment with prednisone, colchicine, and intralesional triamcinolone was initiated. At 4 months of follow-up, the patient had improved.

Key message: First described in 1938, Xanthoma disseminatum also known as Montgomery syndrome is a rare condition. It belongs to the group of non-Langerhans cell histiocytosis that has cutaneous and systemic involvement. Its etiology is unknown, but is considered as a reactive proliferative disorder of histiocytes. It presents at any age with a male predominance, and manifests as cutaneous xanthomas on the face and flexural areas. Mucous membranes, cornea and conjunctive affection are described. Dysphagia and dyspnea are caused by gastrointestinal and respiratory tract involvement. Hypothalamus and pituitary stalk results in diabetes insipidus. Most patients have normal lipids. Histopathology characterizes of a dense dermal infiltrate of histiocytes (S-100-, CD1a-, CD207-, CD68/CD163+, Factor XIIIa+, CD11b+, CD11c+, CD14+, lysozyme+, and alfa-1-antitrypsin+), foamy cells and Toutoun cells. Differential diagnoses include generalized eruptive histiocytoma, multicentric reticulohistiocytosis, Erdheim-Chester disease, Rosai-Dorfman disease, and Langerhans cell histiocytosis. There is not a well-established treatment; steroids, radiation, 2-chorodeoxyadenosine, cyclophosphamide and cyclosporine have been used.